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Case of the Month #169: Septate Uterus with Cervical Duplication and Vaginal Septum

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Clinical Presentation

A 40-year-old woman with a recent diagnosis of cervical cancer after an abnormal Pap smear presents for cancer staging. Her pelvic magnetic resonance image (MRI) is shown below (Figures 1–4).

Diagnosis

Septate uterus with cervical duplication and longitudinal vaginal septum, stage IIa cervical carcinoma.

Radiologic Findings

A septate uterus with a complete septum arising from the midline fundus, 2 separate cervices, longitudinal septum of the vagina, and a flat external uterine fundal contour are shown in Figures 1 and 2. T2-weighted and gadolinium-enhanced fat-saturated 3D T1-weighted images (Figures 3–5) show a enhancing mass within the left cervix, which extends into the upper aspect of the left vaginal canal. It also appears to extend into the vaginal mucosa anteriorly.

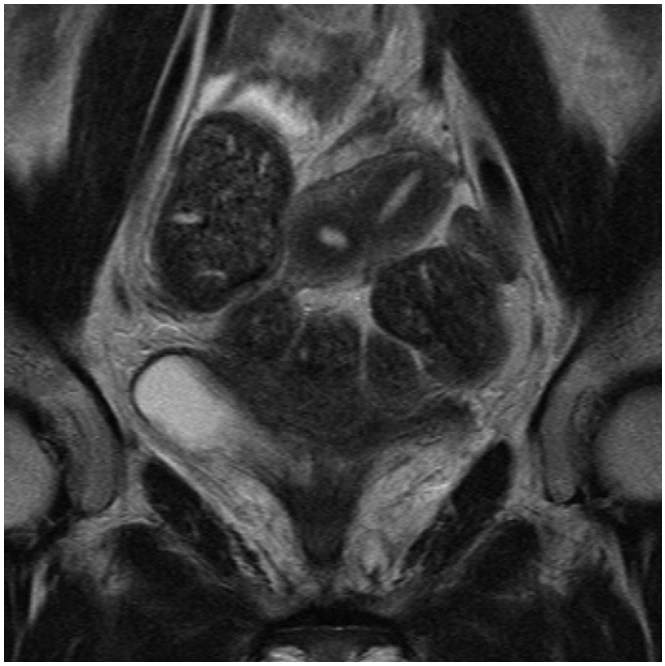

Figure 1. Coronal T2-weighted magnetic resonance image of the pelvis, showing 2 endometrial cavities separated by a septum. The contour of the uterine fundus is flat, with no fundal cleft present.

Discussion

Müllerian duct anomalies have a prevalence that ranges from 0.16%–10% [1]. The cause of most müllerian duct anomalies remains unclear, and the majority of cases are considered to be sporadic or multifactorial [1]. Extrauterine and intrauterine environmental factors, such as exposure to ionizing radiation, intrauterine infections, and teratogenic drugs, such as thalidomide and diethylstilbestrol (DES), have all been shown to cause defects of the developing fetal genital tracts [2].

In embryogenesis, the uterus, fallopian tubes, cervix, and upper two-thirds of the vagina are formed from the 2 müllerian ducts, whereas the lower third of the vagina is developed from the ascending sinovaginal bulb. In general, complete formation of the genital tract is dependent on 3 stages: organogenesis, fusion, and septal resorption [1]. Thus, müllerian duct anomalies are thought to be caused by the failure of the development or fusion of the müllerian ducts and/or the resorption of the septum [3]. The 1988

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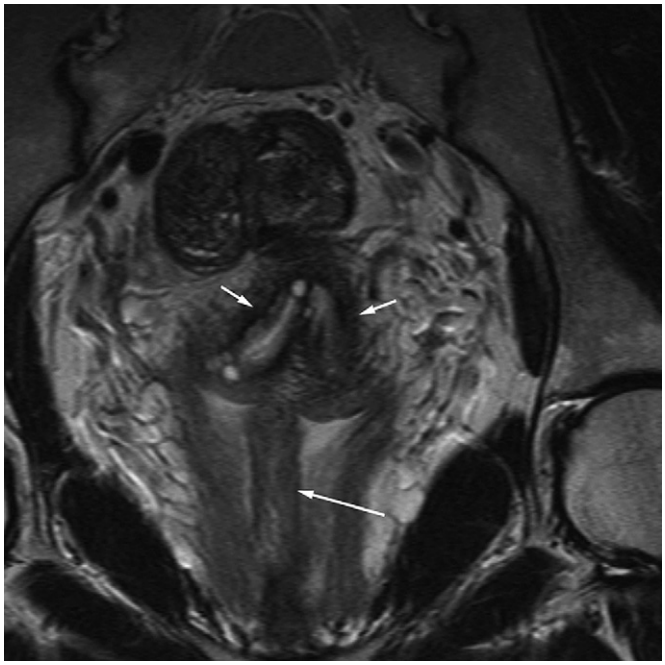

Figure 2. Coronal T2-weighted magnetic resonance image of the pelvis, demonstrating 2 separate cervices (short arrows) and a thick, longitudinal vaginal septum (long arrow).

American Fertility Society (AFS) classification of müllerian anomalies groups müllerian duct anomalies into 7 classes: segmental, unicornuate, didelphus, bicornuate, septate, acrcuate, and DES related [4]. The AFS classification does not include any associated vaginal, urinary, or skeletal abnormalities (Table 1).

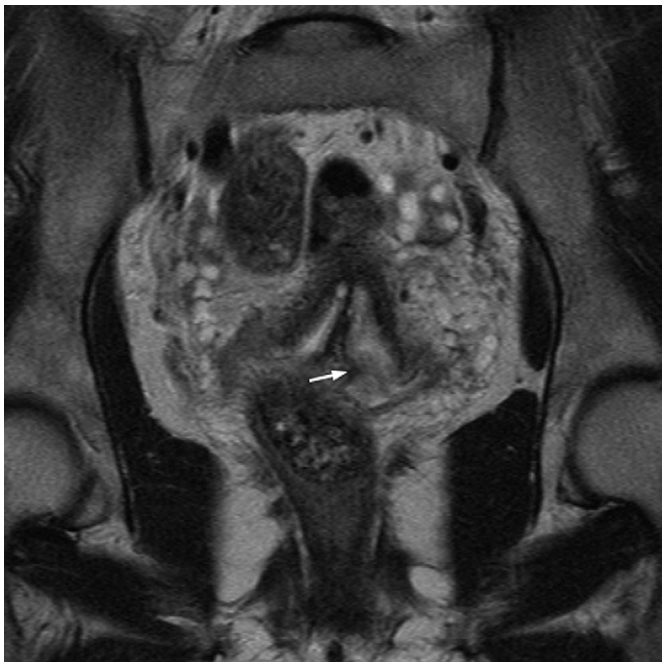

Figure 3. Coronal T2-weighted magnetic resonance image of the pelvis, again demonstrating 2 cervices. An intermediate signal, lobulated mass is seen at the inferior aspect of the left-sided cervix, extending through the os (arrow).

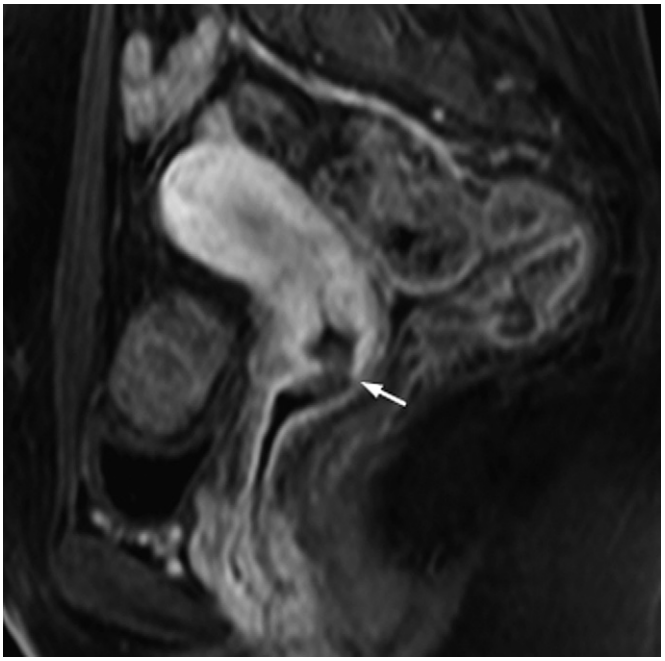

Figure 4. Sagittal contrast-enhanced fat-saturated 3D T1-weighted magnetic resonance images of the pelvis. A polypoid mass (arrow) is seen within the internal os of the left-sided cervix. The mass can be seen protruding into the upper vagina, and invading the anterior vaginal mucosa.

The clinical presentation of a müllerian duct anomaly varies widely, depending on the patient's age and the type of anomaly. Patients may present with primary amenorrhea, an intra-abdominal mass (secondary to hematocolpos or mucocolpos), and cyclical abdominal pain [5]. Although the majority of women with müllerian duct anomalies have little difficulty in conceiving, they are at higher risk of spontaneous abortion, premature delivery, abnormal fetal lie, and dystocia at delivery [2,6].

When müllerian duct anomalies are suspected, a pelvic ultrasound is usually the first examination ordered. More definitive diagnostic modalities include laparoscopic-assisted hysteroscopy, hysterosalpingography and MRI. The role of imaging is to detect, classify, and guide surgical management. MRI is considered the modality of choice for imaging uterine anomalies, because it provides high-resolution images of uterus, including the external uterine contour [1]. MRI has a reported accuracy of up to 100% in the evaluation of müllerian duct anomalies [7–9]. Moreover, it allows the evaluation of the urinary tract for concomitant anomalies [1].

Treatment for müllerian duct anomalies is variable and depends on the type of the anomaly, its clinical manifestations, and the patient's wishes. Operative laparoscopy, microsurgical techniques, and advanced reproductive technologies are now available to minimize operative morbidity and optimize reproductive outcome [10].

In this case, we report a case of müllerian duct anomaly that consists of cervical duplication, longitudinal vaginal septum, uterine septum, and flat fundus. This unique type of müllerian duct anomaly does not fall into the AFS

Table 1
American Fertility Society classification scheme for müllerian duct anomalies[a]

Müllerian duct anomaly class	Description
I: Hypoplasia/agenesis	Includes uterine and/or cervical agenesis or hypoplasia.
II: Unicornuate uterus	Caused by complete, or almost complete, arrest of development of 1 müllerian duct. A rudimentary horn is often present.
III: Didelphys uterus	Secondary to complete nonfusion of 2 müllerian ducts. Individual horns demonstrate complete development and are nearly normal in size. Two cervices are always present.
IV: Bicornuate uterus	Caused by partial nonfusion of the müllerian ducts. There is partial fusion between the 2 horns, which are not fully developed. The lower uterine segment is fused, and the upper segments are separated. Fundal cleft is present.
V: Septate uterus	Septum between the 2 uterine horns fails to be completely resorbed. The septum can be partial or complete. The uterine fundus is typically convex but may be flat or slightly concave.
VI: Arcuate uterus	Single uterine cavity with a convex or flat uterine fundus, but the endometrial cavity demonstrates a small fundal cleft or impression.
VII: DES-related anomaly	May be present in offspring of women exposed to DES during pregnancy. Encompasses various abnormal findings, including uterine hypoplasia, T-shaped uterine cavity, or cavity irregularity.

DES = diethylstilbestrol.
[a] From Ref. 3.

classification system. It does not qualify for septate uterus, because of the presence of cervical duplication, nor does it classify as uterine didelphys, because of the lack of a fundal cleft. According to a MEDLINE search, there have only been a few cases of this anomaly reported in the medical literature to date [4,11–13]. The true incidence of this specific anomaly might be greater than reported, because clinicians are inclined to diagnose a duplicated cervix as uterine didelphys and, therefore, may forgo any further diagnostic workup [3].

The findings of a septate uterus with a cervical duplication and a longitudinal vaginal septum calls into question the classic hypothesis of unidirectional (caudal to cranial) regression of the septum in the uterovaginal canal, where the uterus is initially bicornuate in configuration [14]. Rather, it supports an alternative bidirectional hypothesis proposed by Muller et al [15], which states that the process of fusion and resorption begins at the isthmus, and proceeds simultaneously in both the cranial and caudal directions.

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